Multiple filum terminale hemangioblastomas symptomatic during pregnancy

Case report

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Hemangioblastomas are low-grade, highly vascular tumors commonly associated with von Hippel–Lindau (VHL) syndrome and most often occurring in the cerebellum. Between 3 and 13% of central nervous system hemangioblastomas occur in the spinal cord, and in this case they are usually intramedullary, affecting the posterior aspect of the cord, and superficial or even exophytic. They very rarely occur in spinal nerve roots and if so, they have been reported almost exclusively as part of the VHL syndrome. Hemangioblastomas originating in the filum terminale are exceptional, as demonstrated by the fact that a literature search revealed just nine cases of hemangioblastomas arising in this location, and only four of them were documented on MR images (Table 1). We describe an exceptional case of multiple hemangioblastomas of the filum terminale in a patient with no other evidence of VHL syndrome, in whom pregnancy precipitated symptoms.

**Key Words** • filum terminale hemangioblastoma • multiple spinal tumors • pregnancy • spinal hemangioblastoma • von Hippel–Lindau syndrome

**Case Report**

History and Examination. This 41-year-old woman was in good health until the first trimester of her pregnancy, when she began to experience lumbosacral pain that eventually was relieved only with morphine. Her medical history was unremarkable except that she had had five spontaneous miscarriages, all during the first trimester of pregnancy. She had one healthy child.

The pain, well tolerated at first, increased progressively until the 8th week of gestation when it became excruciating. It was worse when she was in the recumbent position and was associated with lancinating electric-like discharges along the legs and down to the knees. On physical examination we found only generalized hyperreflexia without other signs of pyramidalism. Her muscular balance was Medical Research Council Grade 5/5 and no sensory deficits were seen. The pain was so intense that it prevented walking. Sphincter control was good.

An MR image of the lumbosacral region revealed prominent intradural vessels above the S-1 level associated with two mass lesions at the L-3 and S-1 levels (Fig. 1). The
Masses were isointense compared with the spinal cord on T1-weighted images and moderately hyperintense on T2-weighted images. Gadolinium contrast was not administered because of the pregnancy of the patient. A spinal angiogram to confirm the diagnosis was recommended and was performed after the patient underwent an abortion at another hospital. The angiogram showed a tortuous and dilated artery arising at the T-9 level on the left side, which filled a dense hypervascular tumor at the L-3 level. Large and tortuous veins, running both cephalad and caudally, drained the tumor. The nodule at the S-1 level was fed by small vessels arising from the right hypogastric artery (Fig. 2).

Interestingly, right after the abortion, her pain diminished in a very remarkable way such that common analgesics were enough to control it.

The preoperative diagnosis was multiple hemangioblastomas of the cauda equina. Before the operation, we obtained brain and complete spinal cord MR images that ruled out any other lesion. The patient’s erythrocyte count was within normal range. Results of ophthalmological exploration, abdominal ultrasonography, and genetic workup for VHL syndrome (looking for both punctual alterations and major deletions of the VHL gene) were also normal. The patient’s family was unremarkable for VHL stigmata.

**Operation.** Surgery was performed 2.5 weeks after the pregnancy was terminated. We performed L-3 and S-1 laminectomies and removed en bloc two lesions approximately 2.5 and 3 cm in diameter (Fig. 3). No intraoperative electrophysiological monitoring was performed. The lesions arose from the filum terminale and showed prominent vessel involvement. Results of ophthalmological exploration, abdominal ultrasonography, and genetic workup for VHL syndrome were normal.

**Table 1**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Symptoms</th>
<th>VHL</th>
<th>Lesion Level</th>
<th>MR Imaging</th>
<th>Angiography</th>
<th>Treatment &amp; Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wyburn-Mason, 1943</td>
<td>25, F</td>
<td>radicular pain L2–S2</td>
<td>NR</td>
<td>L2–S2</td>
<td>NA</td>
<td>NR</td>
<td>surgery, recovery</td>
</tr>
<tr>
<td>Norstrom et al., 1961</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td>NR</td>
<td></td>
</tr>
<tr>
<td>Sloof et al., 1964</td>
<td>51, M</td>
<td>NR</td>
<td>no</td>
<td>L4–S1</td>
<td>NA</td>
<td>NR</td>
<td>surgery, outcome NR</td>
</tr>
<tr>
<td>Wolbers et al., 1985</td>
<td>36, M</td>
<td>radicular &amp; lumbar pain, partial cauda equina syndrome back &amp; leg pain, spincter disturbances, partial cauda equina syndrome</td>
<td>no</td>
<td>L-1</td>
<td>NA, diagnoses made by CT</td>
<td>NA</td>
<td>surgery, recovery</td>
</tr>
<tr>
<td>Silverman et al., 1986</td>
<td>66, M</td>
<td>NR</td>
<td>L-2</td>
<td>NA, diagnosis based on lumbar myelogram</td>
<td>NA</td>
<td>NR</td>
<td>surgery, improved symptoms</td>
</tr>
<tr>
<td>Tibbs et al., 1999</td>
<td>35, M</td>
<td>lumbar pain, worse when sitting</td>
<td>no</td>
<td>L2–3</td>
<td>enhancing mass w/ tortuous vessels</td>
<td>NA</td>
<td>surgery, recovery</td>
</tr>
<tr>
<td>Arbelaez et al., 1999</td>
<td>73, M</td>
<td>lumbar &amp; radicular pain</td>
<td>no</td>
<td>L1–2</td>
<td>enhancing mass w/ tortuous vessels</td>
<td>NA</td>
<td>surgery, recovery</td>
</tr>
<tr>
<td>Farnetti et al., 2001</td>
<td>57, M</td>
<td>lumbar &amp; radicular pain, worse while recumbent</td>
<td>no</td>
<td>L-4</td>
<td>enhancing mass w/ tortuous vessels</td>
<td>NA</td>
<td>surgery, recovery</td>
</tr>
<tr>
<td>Nadkami et al., 2006</td>
<td>52, M</td>
<td>lumbar &amp; radicular pain, worse while recumbent, spincter disturbances</td>
<td>no</td>
<td>L2–3</td>
<td>enhancing mass w/ tortuous vessels</td>
<td>yes</td>
<td>surgery w/ preop embolization, recovery</td>
</tr>
<tr>
<td>present case</td>
<td>41, F</td>
<td>lumbar pain, worse while recumbent, better after abortion</td>
<td>no</td>
<td>L-3 &amp; S-1 (two lesions)</td>
<td>iso- &amp; hyperintense masses w/ tortuous vessels, no contrast</td>
<td>yes</td>
<td>surgery, recovery</td>
</tr>
</tbody>
</table>

* NA = not obtained; NR = not reported.
nent arterial and venous vessels on their surface that were coagulated and cut together with the filum. The lesions were adherent to the cauda equina nerve roots, which were carefully spared except for a tiny root that was totally embedded by the caudal tumor and thus was sacrificed.

**Postoperative Course.** After the operation, the patient was free of pain and showed no neurological deficits except for a slight hypesthesia in the right perineal area. One year later she is asymptomatic except for the aforementioned sensory deficit.

Pathological examination of the two masses showed large epithelioid stromal cells with vacuolated cytoplasm and a small nucleus in a rich capillary network with several larger vessels (Fig. 4). No atypias were found. The histological diagnosis was hemangioblastoma in both cases.

**Discussion**

Spinal cord hemangioblastomas are infrequent tumors that comprise between 1.6 and 2.1% of all spinal cord tumors. Extramedullary hemangioblastomas are even more rare, accounting for only one fifth of all spinal cord hemangioblastomas. Approximately 25% of these tumors are associated with VHL syndrome, but it has been suggested that this estimate may be artificially low because some patients with hemangioblastoma do not undergo appropriate screening for VHL syndrome. Patients with hemangioblastoma associated with VHL syndrome are younger and more frequently harbor spinal hemangioblastomas than patients with sporadic tumors. Multiplicity is seen almost exclusively in VHL disease.

The present case exhibits two very rare features that make it exceptional. The first is the presence of multiple extramedullary hemangioblastomas arising in the filum terminale, even in the absence of any sign of VHL syndrome, and the second is the relationship of the symptoms with the evolution of pregnancy in the patient.

To the best of our knowledge, only nine cases of hemangioblastomas arising in the filum terminale have been reported (Table 1).
documented on MR images. None of the cases included multiple tumors. The case we describe seems to be the first report of multiple hemangioblastomas arising in the filum terminale. Curiously, no patient with hemangioblastomas in the filum terminale has had the VHL syndrome, and neither did our patient despite the fact that she had multiple extramedullary hemangioblastomas. Although the presence of more than one hemangioblastoma is one of the clinical criteria to justify the diagnosis of VHL syndrome, in the present case neither a family history of VHL disease nor any other features of the disease, including irregularities on genetic analysis, were present. Because the detection rate for VHL mutations is nearly 100% in genetic studies in VHL syndrome, it is very probable that our patient did not harbor the VHL disease.

With regard to the natural history of hemangioblastomas, it has been reported that pregnancy seems to hasten the symptoms caused by these lesions. Most authors consider this factor to be related to an increase in the volume of the intravascular compartment, leading to an augmentation in tumor size. Endocrinological changes may also play a role, as demonstrated by the presence of progesterone receptors in some of these tumors. The majority of the reports dealing with the relationship between hemangioblastomas and pregnancy are focused on cerebellar hemangioblastomas. In some of these cases an improvement in symptoms after delivery has been described. Given the rarity of spinal hemangioblastomas, their biological behavior in pregnant women is not well understood. We found only two reports relating spinal hemangioblastomas and gestation. The first case, reported by Ogasawara et al., features the acute onset during pregnancy of paraplegia due to intramedullary hemorrhage in a patient with a hemangioblastoma. The second case, described by Kurne et al., features a patient harboring several spinal and cerebellar hemangioblastomas with fluctuating spinal cord symptoms during her second and third pregnancies. Her clinical status worsened during the second pregnancy, improved after delivery, and recurred during the third pregnancy. This behavior is similar to that described in the present case.

![Figure 3](image1.png)

**Fig. 3.** Photographs depicting surgically excised specimens of intradural lumbar (upper) and sacral (lower) hemangioblastomas. The tumors are smooth and lobulated with large and dilated vessels in their surfaces.

![Figure 4](image2.png)

**Fig. 4.** Photomicrographs demonstrating neuropathological findings. *Left:* Tumor at the S-1 level. Note the fine vascular capsule (arrow) and the rich capillary network. *Right:* Tumor at the L-3 level. Note the large cells with vacuolated cytoplasm and small nucleus. H & E, original magnification × 10 (left) and × 40 (right).
which showed a dramatic relationship between symptoms and the evolution and termination of pregnancy.

Conclusions

In summary, we presented the case of a patient with two hemangioblastomas arising in the filum terminale but without any other sign of VHL disease. The lesions became symptomatic during her pregnancy, followed by a rapid alleviation of the symptoms on termination of the pregnancy. The coexistence of such features makes this an exceptional case that clearly exemplifies the relationship between spinal vascular tumors and pregnancy.

References


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